

Anesthesia for correction of esophageal stricture in a patient with epidermolysis bullosa: a case report

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Introduction

Epidermolysis bullosa dystrophica (EBD), first described by Fox in 1879, is an autosomal recessive disorder with an incidence of 1 in 300 000 births [1]. The disorder is characterized by bullae and blister formation on the skin that may occur spontaneously or as a result of minor trauma. In addition to the skin, the mucous membranes of the mouth, pharynx, esophagus, and anus are commonly affected. Recurrent blistering lesions of the esophagus may occur, which may lead to esophageal stenosis and difficulty in maintaining adequate nutrition. The combination of inadequate nutrition and the constant loss of fluid, protein, and blood from the various lesions results in malnutrition, dehydration, electrolyte disturbances, hypoproteinemia, and anemia.

EBD is an important disease from the anesthetist's point of view because of the unfortunate sequelae that can occur if the necessary precautions are not taken. Anemia, hypoproteinemia, and electrolyte imbalance frequently demand preoperative correction. Intubation in EBD can be difficult because of mouth contractures and poor dentition, both of which can impede laryngoscopy. Because trauma from adhesive tape, blood pressure cuffs, tourniquets, or adhesive ECG electrodes may cause the formation of bullae, anesthetic management of patients with EBD is directed toward avoiding even minor trauma to the skin and mucous membranes.

There have been several reports concerning the anesthetic management of patients with EBD for various

different surgical procedures. We could find only two cases of colonic interposition for esophageal stricture with EBD [2,3]. The anesthetic methods were different, and the procedures were shorter than those used in our case. There was only one patient with EBD who underwent a reconstructive operation of 12h duration [4]. Because of the paucity of the literature on anesthesia of long duration in patients with EBD, we report the anesthetic technique employed in a 12-year-old girl with EBD who underwent a colonic interposition operation of more than 10h duration.

Case report

The patient was a 12-year-old-girl, 125cm tall and weighing 24kg. She had a history of EBD since birth, and she was admitted for investigation of vomiting. A mid-esophageal stricture was shown by barium esophagogram. On physical examination, her body surface was covered with numerous blisters and with fresh and scattered bullae, particularly involving the face, the extensor sides of the extremities, and around the mouth and nose. There was evidence of microstomia, poor dentition, and limited temporomandibular joint mobility. There were also flexion contractures of the fingers of both hands (Fig. 1). The preoperative hemoglobin level was 10.3 g·dl⁻¹. Serum electrolytes, protein, and blood urea nitrogen were within normal limits. She had been treated with corticosteroids in the past, but at the time of admission she was taking phenytoin and was applying hydrocortisone ointment 0.5% to her lesions.

The patient was taken to the operating room without premedication, and she placed herself on the operating table, which was fitted with cotton rolls. Anesthesia was induced with vital capacity inhalation of sevoflurane in combination with nitrous oxide and oxygen by a disposable face mask. A 25G intravenous cannula was inserted in the right forearm, and an

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Fig. 1. Typical appearance of body surface of patient with epidermolysis bullosa dystrophica

intraarterial cannula was inserted in the left radial artery for monitoring of blood pressure and for blood gas analyses. Both catheters were sutured to the skin. A central venous catheter was then inserted into the right subclavian vein and again sutured to the skin. A pulse oximeter probe was positioned on the earlobe, and wet pad electrodes were lightly applied for electrocardiographic (ECG) monitoring. After the administration of succinylcholine 25mg intravenously, tracheal intubation was gently performed with a well-lubricated laryngoscope and a 5-mm-cuffed orotracheal tube. The endotracheal tube was secured with surgical drape. Her eyes were lubricated and protected with wet gauze (Fig. 2).

Anesthesia was maintained by intermittent positive-pressure ventilation using sevoflurane (1%–2%), and nitrous oxide in oxygen (50%). Intermittent doses of vecuronium (10mg total) were given for neuromuscular relaxation, and intermittent doses of alfentanil (1000µg total) for analgesia. Intravenous fluids consisted of 5% dextrose in 0.2% sodium chloride (1500ml total) and 400ml of transfused blood to replace the loss. The surgical procedure consisted of right colonic interposition to bypass the esophageal stricture and gastrostomy via a midline abdominal incision. During the operation, the arterial blood pressure, arterial blood gases, SpO₂, and heart rate were stable and showed no significant changes, compared with preanesthetic values. The positions of the patient's head, the upper extremities, and the pulse oximeter were changed every hour. The total anesthesia time was 10h 30min. At the end of the operation, neostigmine (0.05mg·kg⁻¹) and atropine



Fig. 2. Use of surgical drape for security of endotracheal tube and protection of eyes with wet gauze

(0.01mg·kg⁻¹) were administered and the trachea was extubated. The preoperative and postoperative courses were uneventful. However, although all possible precautions had been taken, several bullae developed around the mouth and incision sites.

Discussion

A review of the anesthesia literature shows that several case reports about the anesthetic management of patients with EBD have been documented, but until now there have been only two patients who underwent colonic interposition for esophageal stricture [2,3] (Table 1). The total duration of anesthesia in these two cases was shorter than that in our case. Yonker-Sell and Connolly [4] reported another patient with EBD who underwent 12h of anesthesia for a free flap to the right hand with release of syndactyly. They used ketamine for induction and isoflurane and continuous fentanyl infusion for maintenance. A variety of anesthetic techniques have been described in patients with EBD. Ketamine anesthesia has been used for patients with epidermolysis bullosa [1,5]. Idvall [6] suggested ketamine with the continuous infusion technique for long-lasting procedures. However, ketamine is contraindicated in the presence of hypertension, myocardial insufficiency, psychiatric disorders, or raised intracranial pressure. Also, excitation during recovery may cause further trauma [7]. Thiopentone and propofol have also been used for induction and maintenance of anesthesia [5,7]. Brachial plexus [1], spinal, and epidural [7,8] blocks in EBD patients have been reported. Inhalation induction was the most common method of induction because of difficulty in gaining

Table 1. Two cases of colonic interposition for esophageal stricture in EBD

Reference	Case	Type of operation	Duration of anesthesia	Anesthetic agents	Complications
Tomlinson [2]	61-year-old woman	Colonic interposition for esophageal stricture	5h	Thiopentone, suxamethonium, halothane, alcuronium, papaveretum	Small bullae on electrodes sites
Milne and Rosales [3]	10-year-old girl	Colonic interposition for esophageal stricture	7h	Halothane, N ₂ O/O ₂ , suxamethonium, droperidol, fentanyl, pancuronium	None

EBD, epidermolysis bullosa dystrophica

venous access [5]. Sevoflurane suits these patients with rapid induction and recovery [7]. With sevoflurane, unlike halothane, postoperative involuntary movements and shivering are minimal, with less risk of inadvertent trauma [7]. In this connection, we preferred sevoflurane anesthesia in our case. The hemodynamic situation was stable and awakening was fast and uncomplicated.

The use of neuromuscular blocking agents has previously been questioned [1,7]. It has been suggested that suxamethonium-induced fasciculations could lead to trauma to the body surface and excessive release of potassium in the presence of muscular atrophy [1]. However, suxamethonium has previously been used uneventfully [3,5]. Our patient was successfully intubated after the use of suxamethonium, and no fasciculations occurred. Nondepolarizing muscle relaxants were also used to a lesser degree, because low albumin concentrations are known to change the pharmacokinetics of neuromuscular blockers [1,7]. Both types of neuromuscular relaxant were used in our patient, with no unwanted sequelae.

Ames et al. [7] reported new facial blisters that were caused by the face mask after operation. Numerous fresh bullae were seen around the mouth in our patient after the operation, possibly caused by the use of surgical drape for tube security or application of a face mask in direct contact with the patient's face.

Blood-pressure measurement is essential for major procedures, and intraarterial monitoring has been recommended [5]. Intraarterial and intravenous catheters should be sutured in place or held in place by a gauze wrap. Indirect blood-pressure measurement requires an adequately padded cuff. In our patient, intraarterial monitoring was used because of the long duration of surgery. Formation of new bullae fortunately did not occur at the cannulation sites.

Airway patency may be a problem in patients with EBD because microstomia, poor dentition, strictures, or webbing in the oropharynx can make intubation impossible. Several cases have been reported in which endotracheal intubation was performed successfully

with no sequelae after extubation [2,3]. Nevertheless, others have warned against this risk [5]. If tracheal intubation is necessary, it is important to ensure that both laryngoscopy and intubation are atraumatic. A well-lubricated laryngoscope and endotracheal tube are preferred to avoid trauma. The use of a laryngeal mask airway (LMA) has also been described for patients with EBD. Ames et al. [7] used an LMA in 57 cases, with only one case of new lingual bullae. In our patient, tracheal intubation was an essential part of the anesthetic technique, and there were no problems of upper airway obstruction or stridor after extubation.

As a result of the findings of this and other case reports, prolonged anesthetic management of patients with EBD should include careful airway manipulation, reduction of mucocutaneous contacts, prevention of pressure or friction skin trauma, appropriate electrolyte and plasma volume replacement, and use of nonadhesive material. We believe sevoflurane might be useful for these patients, with rapid induction and recovery.

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